General

Guideline Title

ACG clinical guideline: evidenced based approach to the diagnosis and management of esophageal eosinophilia and eosinophilic esophagitis (EoE).

Bibliographic Source(s)

Dellon ES, Gonsalves N, Hirano I, Furuta GT, Liacouras CA, Katzka DA. ACG clinical guideline: evidenced based approach to the diagnosis and management of esophageal eosinophilia and eosinophilic esophagitis (EoE). Am J Gastroenterol. 2013 May;108(5):679-92. PubMed

Guideline Status

This is the current release of the guideline.

Recommendations

Major Recommendations

The definitions for quality of evidence (strong, moderate, low, and very low) and strength of recommendations (strong or conditional) are provided at the end of the "Major Recommendations" field.

Diagnosis

Definition and Causes of Esophageal Eosinophilia

1. Esophageal eosinophilia, the finding of eosinophils in the squamous epithelium of the esophagus, is abnormal and the underlying cause should be identified (Recommendation strong, evidence moderate).

Definition of Eosinophilic Esophagitis (EoE) and Diagnostic Criteria

2. EoE is a clinicopathologic disorder diagnosed by clinicians taking into consideration both clinical and pathologic information without either of these parameters interpreted in isolation, and defined by the following criteria:
    - Symptoms related to esophageal dysfunction
    - Eosinophil-predominant inflammation on esophageal biopsy, characteristically consisting of a peak value of ≥15 eosinophils per high-power field (eos/hpf)
    - Mucosal eosinophilia is isolated to the esophagus and persists after a proton-pump inhibitor (PPI) trial
    - Secondary causes of esophageal eosinophilia excluded (see Table 2 in the original guideline document)
    - A response to treatment (dietary elimination; topical corticosteroids) supports, but is not required for, diagnosis (Strong recommendation, low evidence).
3. Esophageal biopsies are required to diagnose EoE; 2–4 biopsies should be obtained from both the proximal and distal esophagus to

maximize the likelihood of detecting esophageal eosinophilia in all patients in whom EoE is being considered (Recommendation strong, evidence low).
4. At the time of initial diagnosis, biopsies should be obtained from the antrum and/or duodenum to rule out other causes of esophageal eosinophilia in all children and in adults with gastric or small intestinal symptoms or endoscopic abnormalities (Recommendation strong, evidence low).

Diagnostic Challenges: PPI-Responsive Esophageal Eosinophilia and Gastroesophageal Reflux Disease (GERD)

5. Proton-pump inhibitor-responsive esophageal eosinophilia (PPI-REE) should be diagnosed when patients have esophageal symptoms and histologic findings of esophageal eosinophilia, but demonstrate symptomatic and histologic response to proton-pump inhibition. At this time, the entity is considered distinct from EoE, but not necessarily a manifestation of GERD (Recommendation conditional, evidence low).
6. To exclude PPI-REE, patients with suspected EoE should be given a 2-month course of a PPI followed by endoscopy with biopsies (Recommendation strong, evidence low).
7. A clinical, endoscopic and/or histologic response to a PPI does not establish gastroesophageal reflux as the cause of esophageal eosinophilia. To determine whether reflux is contributing to esophageal eosinophilia, additional evaluation for GERD, as per standard clinical practice, is recommended. This may include ambulatory pH testing in selected cases (Recommendation conditional, evidence low).

Treatment

Endpoints of Treatment in EoE

8. The endpoints of therapy of EoE include improvements in clinical symptoms and esophageal eosinophilic inflammation. While complete resolution of symptoms and pathology is an ideal endpoint, acceptance of a range of reductions in symptoms and histology is a more realistic and practical goal in clinical practice (Recommendation conditional, evidence low).
9. Symptoms are an important parameter of response in EoE, but cannot be used alone as a reliable determinant of disease activity and response to therapy, given that compensatory dietary and lifestyle factors can mask symptoms (Recommendation conditional, evidence moderate).

Pharmacologic Treatments

10. Topical steroids (i.e., fluticasone or budesonide, swallowed rather than inhaled, for an initial duration of 8 weeks) are a first-line pharmacologic therapy for treatment of EoE (Recommendation strong, evidence high).
11. Prednisone may be useful to treat EoE if topical steroids are not effective or in patients who require rapid improvement in symptoms (Recommendation conditional, evidence low).
12. Patients without symptomatic and histologic improvement after topical steroids might benefit from a longer course of topical steroids, higher doses of topical steroids, systemic steroids, elimination diet, or esophageal dilation (Recommendation conditional, evidence low). There are few data to support the use of mast cell stabilizers or leukotriene inhibitors, and biologic therapies remain experimental at this time.

Dietary Treatments

13. Dietary elimination can be considered as an initial therapy in the treatment of EoE in both children and adults (Strong recommendation, evidence moderate).
14. The decision to use a specific dietary approach (elemental, empiric, or targeted elimination diet) should be tailored to individual patient needs and available resources (Recommendation conditional, evidence moderate).
15. Clinical improvement and endoscopy with esophageal biopsy should be used to assess response to dietary treatment when food antigens are either being withdrawn from or reintroduced to the patient (Recommendation conditional, evidence low).
16. Gastroenterologists should consider consultation with an allergist to identify and treat extraesophageal atopic conditions, assist with treatment of EoE, and to help guide elemental and elimination diets (Recommendation conditional, evidence low).

Endoscopic Treatment

17. Esophageal dilation, approached conservatively, may be used as an effective therapy in symptomatic patients with strictures that persist in spite of medical or dietary therapy (Recommendation conditional, evidence moderate).
18. Patients should be well informed of the risks of esophageal dilation in EoE including post-dilation chest pain, which occurs in up to 75 % of patients, bleeding, and esophageal perforation (Recommendation conditional, evidence moderate).

Outcomes

Natural History of EoE

19. While knowledge of the natural history of EoE is limited, patients should be counseled about the high likelihood of symptom recurrence after discontinuing treatment due to the chronic nature of this disease (Recommendation strong, moderate evidence).

Maintenance Therapy

20. The overall goal of maintenance therapy is to minimize symptoms and prevent complications of EoE, preserve quality of life, with minimal long-term adverse effects of treatments (Recommendation conditional, evidence low).
21. Maintenance therapy with topical steroids and/or dietary restriction should be considered for all patients, but particularly in those with severe dysphagia or food impaction, high-grade esophageal stricture and rapid symptomatic/histologic relapse following initial therapy (Recommendation conditional, evidence low).

Definitions:

Quality of Evidence Using the Grading of Recommendations Assessment, Development, and Evaluation (GRADE) System

The quality of evidence was either strong (further research is unlikely to change confidence in the estimate), moderate (further research is likely to change confidence in the estimate), low (further research is very likely to change confidence in the estimate), or very low (the estimate of the effect is very uncertain).

Strength of Recommendations Using the Grade System

Recommendations were either strong (desirable effects outweigh undesirable effects) or conditional (trade-offs are less certain).

Clinical Algorithm(s)

The original guideline document contains a clinical algorithm for the approach to esophageal eosinophilia (EoE) and diagnosis of EoE.

Scope

Disease/Condition(s)

- Esophageal eosinophilia
- Eosinophilic esophagitis (EoE)

Guideline Category

Diagnosis

Evaluation

Management

Treatment

Clinical Specialty

Allergy and Immunology

Family Practice

Gastroenterology

Internal Medicine

Pathology

Pediatrics

Intended Users

Advanced Practice Nurses

Nurses

Physician Assistants

Physicians

Guideline Objective(s)

To provide recommendations for the evaluation and management of patients with esophageal eosinophilia and eosinophilic esophagitis (EoE)

Target Population

Adults and children with esophageal eosinophilia and eosinophilic esophagitis (EoE)

Interventions and Practices Considered

Diagnosis

1. Assessment of signs and symptoms
2. Endoscopy
3. Esophageal biopsies

Management/Treatment

1. Management of proton-pump inhibitor (PPI) responsive esophageal eosinophilia
    - PPI for 2 months, followed by endoscopy with biopsies
2. Assessment of symptom improvement
3. Pharmacologic therapy
    - Topical steroids (fluticasone/budesonide)
    - Prednisone
4. Dietary treatments
    - Elimination diet
5. Consultation with allergist
    - Identification and treatment of extra-esophageal atopic conditions
6. Endoscopic treatment
    - Esophageal dilation
7. Maintenance therapy
    - Topical steroids and/or dietary restriction

Major Outcomes Considered

- Predictive/ prognostic value of diagnostic tests
- Efficacy of treatment
- Symptom control
- Adherence to treatment regimen
- Incidence of symptom recurrence after discontinuing treatment
- Quality of life

Methodology

Methods Used to Collect/Select the Evidence

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

A search of PubMed and Ovid was performed using the search terms 'eosinophilic esophagitis', 'esophageal eosinophilia', 'ringed esophagus' and 'esophagus eosinophils' for years 1993 to present. There were no specific inclusion or exclusion criteria.

Number of Source Documents

Not stated

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

The Grading of Recommendations Assessment, Development, and Evaluation (GRADE) system was used to evaluate the quality of supporting evidence. The quality of evidence was either strong (further research is unlikely to change confidence in the estimate), moderate (further research is likely to change confidence in the estimate), low (further research is very likely to change confidence in the estimate), or very low (the estimate of the effect is very uncertain).

Methods Used to Analyze the Evidence

Review of Published Meta-Analyses

Systematic Review

Description of the Methods Used to Analyze the Evidence

See the "Rating Scheme for the Strength of the Evidence" field.

Methods Used to Formulate the Recommendations

Expert Consensus

Description of Methods Used to Formulate the Recommendations

See the "Rating Scheme for the Strength of the Recommendations" field.

Rating Scheme for the Strength of the Recommendations

The Grading of Recommendations Assessment, Development, and Evaluation (GRADE) system was used to grade the strength of recommendations. Recommendations were either strong (desirable effects outweigh undesirable effects) or conditional (trade-offs are less certain).

Cost Analysis

A cost analysis was not performed and published cost analyses were not reviewed.

Method of Guideline Validation

Internal Peer Review

Description of Method of Guideline Validation

In an effort to make our new guidelines as "fresh" as possible when published, we have created a special guideline review process, involving members of the Board of Trustees, Practice Parameters Committee and the American Journal of Gastroenterology. It is our goal to review the guideline, allow you to revise the guideline, and re-review the guideline within 6 months of first submission. Therefore the entire process should take 1 year from commission to finished, accepted guideline.

Evidence Supporting the Recommendations

Type of Evidence Supporting the Recommendations

The type of supporting evidence is identified and graded for each recommendation (see the "Major Recommendations" field).

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

Appropriate diagnosis and management of patients with esophageal eosinophilia and eosinophilic esophagitis (EoE)

Potential Harms

Risks of Esophageal Dilation

- Patients should be well informed of the risks of esophageal dilation in esophageal eosinophilia (EoE) including post-dilation chest pain, which occurs in up to 75 % of patients, bleeding, and esophageal perforation.
- In two recent meta-analyses of esophageal dilation in EoE, the perforation rate for dilation was 0.3 %, similar to that quoted for esophageal dilation for other benign esophageal diseases. It is important to note, however, that this rate was achieved at academic centers experienced in performing esophageal dilation in patients with EoE.
- The most common risk of dilation in EoE is post-procedural chest pain, reported in almost three quarters of patients when asked about this symptom prospectively. Major bleeding defined by need for endoscopic hemostasis or blood product is rare (only 1 patient reported to date).

Qualifying Statements

Qualifying Statements

The majority of recommendations in the original guideline document are "conditional" rather than "strong", further emphasizing the paucity of firm data guiding decisions and the likelihood of changing consensus in answer to even some of the most basic questions about this disease.

Implementation of the Guideline

Description of Implementation Strategy

An implementation strategy was not provided.

Implementation Tools

Clinical Algorithm

Patient Resources

For information about availability, see the *Availability of Companion Documents* and *Patient Resources* fields below.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

Getting Better

Living with Illness

IOM Domain

Effectiveness

Patient-centeredness

Identifying Information and Availability

Bibliographic Source(s)

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Adaptation

Not applicable: The guideline was not adapted from another source.

Date Released

2013 May

Guideline Developer(s)

American College of Gastroenterology - Medical Specialty Society

Source(s) of Funding

American College of Gastroenterology

Guideline Committee

Not stated

Composition of Group That Authored the Guideline

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Financial Disclosures/Conflicts of Interest

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Potential Competing Interests

David A. Katzka, Nirmala Gonsalves declare no conflict of interest. Evan S. Dellon is a consultant to Oncoscope. Christopher A. Liacouras is a speaker for Nutricia, and is on the American Partnership for Eosinophilic Disorders. Ikuo Hirano is on the advisory board of Meritage and Aptalis.

Guideline Status

This is the current release of the guideline.

Guideline Availability

Electronic copies: Available from the American College of Gastroenterology (ACG) Web site .

Availability of Companion Documents

The following is available:

- American College of Gastroenterology Practice Parameters Committee. Guideline development policies. 2010 Jan. Available from the American College of Gastroenterology (ACG) Web site .

Patient Resources

Information on eosinophilic esophagitis (EoE) is available on the American College of Gastroenterology's Patient Education & Resource Center Web site .

Please note: This patient information is intended to provide health professionals with information to share with their patients to help them better

understand their health and their diagnosed disorders. By providing access to this patient information, it is not the intention of NGC to provide specific medical advice for particular patients. Rather we urge patients and their representatives to review this material and then to consult with a licensed health professional for evaluation of treatment options suitable for them as well as for diagnosis and answers to their personal medical questions. This patient information has been derived and prepared from a guideline for health care professionals included on NGC by the authors or publishers of that original guideline. The patient information is not reviewed by NGC to establish whether or not it accurately reflects the original guideline's content.

NGC Status

This NGC summary was completed by ECRI Institute on June 20, 2013. The information was verified by the guideline developer on June 25, 2013.

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